

Laparoscopic Duodenojejunostomy for Superior Mesenteric Artery Syndrome

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ABSTRACT

Background: Superior mesenteric artery (SMA) syndrome, also called Wilkie's syndrome, is a rare clinical phenomenon believed to be caused by compression of the third portion of the duodenum by the overlying superior mesenteric artery. We present the case of a 32-year-old female who presented with epigastric pain, weight loss, and vomiting.

Methods: Her workup included a normal upper endoscopy as well as an abdominal CT scan and upper GI contrast study that confirmed the diagnosis of superior mesenteric artery syndrome. The patient was taken to the operating room and underwent successful treatment with laparoscopic duodenojejunostomy.

Results: The patient achieved complete relief of her symptoms and is able to eat a regular diet without difficulty. SMA syndrome is a real anatomic clinical pathology resulting in chronic, consistent obstructive symptoms. An upper GI series and CT scan with contrast can confirm the diagnosis.

Conclusion: Laparoscopic duodenojejunostomy should be considered the treatment of choice for these patients, because it offers a high likelihood of excellent outcome based on the current literature.

Key Words: Superior mesenteric artery syndrome, Duodenojejunostomy, Laparoscopy.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome, also called Wilkie's syndrome, is a rare clinical phenomenon believed to be caused by compression of the third portion of the duodenum by the overlying superior mesenteric artery. An abnormally acute aorta-SMA angle, or high retroperitoneal attachment of the ligament of Treitz, or both, creates duodenal compression in SMA syndrome. This phenomenon has been recognized as a named clinical entity after its original description in 1861. The incidence of duodenal compression within the aorto-SMA angle has been estimated to be as high as 0.3% from upper gastrointestinal barium swallow studies,¹ but the incidence of clinically significant and appropriately confirmed disease has been estimated to be much lower (range, 0.01–0.08%).^{1,2} Acceptance of this cause as a clinically reversible pathology has been controversial,³ but recent evolution of the literature seems to be clarifying the issue. Medically refractory cases have in the past been treated by open release of the ligament of Treitz, allowing mobilization of the duodenum or by enteric bypass. As we move into the minimally invasive era, operative intervention for the disease can and should be handled via the laparoscopic approach. In this case, we describe laparoscopic duodenojejunostomy to successfully treat SMA syndrome and review the literature on the subject.

CASE REPORT

Presentation

A 32-year-old woman presented with a 5-year history of vague epigastric pain that intermittently radiated to the back. She had at that time been found on 2 occasions to have elevated pancreatic enzymes and was treated conservatively for pancreatitis. She lost 10 pounds to 20 pounds after each of these episodes per history. Extensive workup at that time including CT scan, MRI, and ERCP revealed no identifiable anatomic abnormalities. At the time of presentation to our institution, 3 years had passed, and the patient had persistent chronic pain and progressively worsening nausea. Over the previous 6 months, she had lost another 30 pounds. Her epigastric pain increased

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with meals, and she subsequently developed profuse vomiting after large meals without antecedent symptoms. In an attempt to alleviate these symptoms, she would eat smaller meals and consume fewer liquids, which led to dehydration, fatigue, and constipation. A CT scan revealed a 4-cm dilated duodenal bulb with abrupt decompression to normal duodenum beyond the SMA (**Figure 1**). The distance between the SMA and aorta at the level of the duodenal lumen was 7.2 mm. An upper gastrointestinal swallow study revealed a dilated proximal duodenum with an abrupt vertical cutoff (**Figure 2**). During fluoroscopy, there was significant “to-and-fro” peristalsis of the second and third portions of the duodenum against an apparent obstruction, with small jets of contrast squirting through. After contrast went through, a narrow lumen at the point of obstruction with distal decompressed bowel could be seen (**Figure 3**). Upper endoscopy revealed no intraluminal pathology. The diagnosis of SMA syndrome was assumed, and the patient was counseled on laparoscopic duodenojejunostomy.

Operation

At operation, the patient was placed in the supine position with both arms tucked. Insufflation was attained by using an insufflation needle (Ethicon Pneumoneedle, Ethicon Endo-Surgery Inc., Cincinnati, OH) through a 10-mm supraumbilical curvilinear incision. A 10-mm Optiview port

(Ethicon Non-bladed Trocar 10/12, Ethicon Endo-Surgery, Cincinnati, OH) containing a 10-mm, 0-degree scope with the camera focused to the tip of the port was used to enter the abdomen under direct visualization. A 12-mm port was then placed in the left mid subcostal region with a 5-mm port between the two. A 5-mm port was then placed in the right mid subcostal region. The camera was then switched for a 10-mm, 30-degree lens, and the handle was attached to the robotic endoscopic positioning system for audio-activated control (AESOP, Computer Motion Co., Sunnyvale, CA).

Omentum and transverse colon were retracted cephalad, revealing the large dilated duodenum bulging through a thin, attenuated mesocolon, which was incised to expose the surface of the duodenum. The ligament of Treitz was identified, and the proximal jejunum was run approximately 20cm to the first loop that could be brought easily to the dilated duodenum without tension. Utilizing a 2–0 Surgidac suture on the laparoscopic stitching device (USSC Endostitch 173016, US Surgical Corp., Norwalk, CT), a running suture was placed to secure the 2 limbs of bowel and serve as the back row in preparation for a 2-layer, side-to-side anastomosis.

The duodenum and jejunum were entered with the Harmonic shears (Ultracision Harmonic Scalpel, Ethicon Endo-Surgery, Inc., Cincinnati, OH) approximately 3 mm

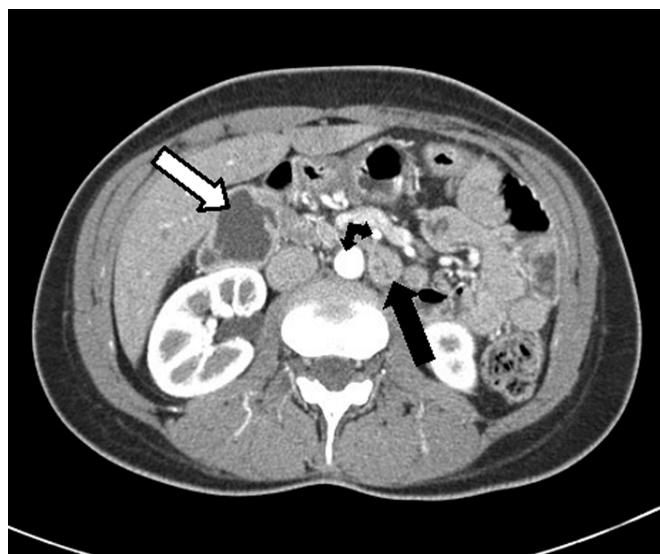


Figure 1. Computed tomography of the abdomen revealed dilated proximal duodenum (large white arrow), decompressed distal duodenum (large black arrow), and narrow distance between the superior mesenteric artery and aorta (small double arrow).

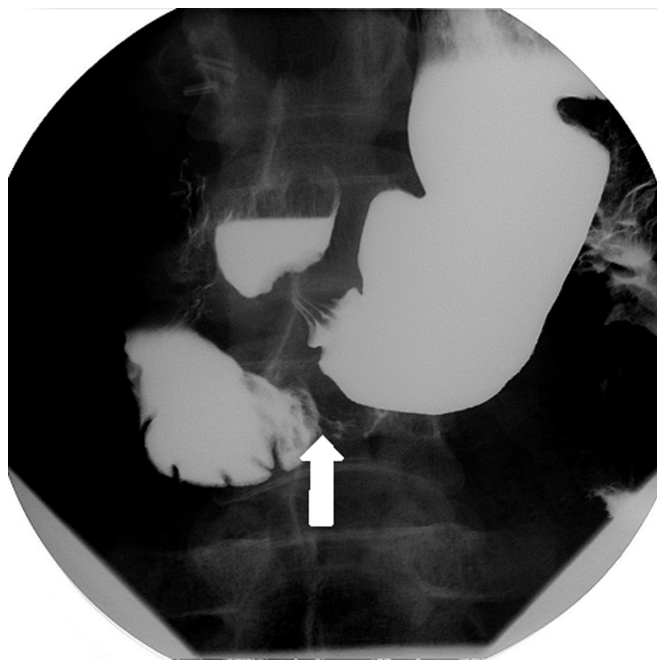


Figure 2. Contrast study before surgery demonstrating dilated proximal duodenum with a sharp vertical cutoff (arrow).

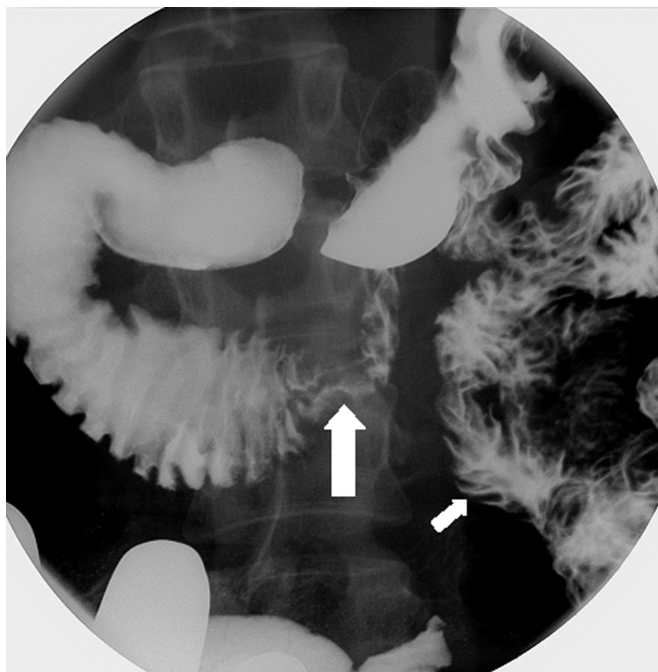


Figure 3. Delayed images showing narrowed duodenal lumen at the level of obstruction (large arrow) with distal decompression (small arrow).

above the posterior row, and the stapling device (2.5 mm/45 mm) was deployed. The defect was closed with running absorbable suture using the suture device to complete the inner layer of the anastomosis. The outer layer was then completed with a running nonabsorbable stitch joining the ends of the originally placed posterior row.

One intraperitoneal drain was left at the anastomosis and brought out through the right subcostal port site.

Outcome

A swallow study performed on postoperative day 1 revealed no evidence of leakage or stenosis, and the patient was started on a clear liquid diet and advanced to pureed foods over the next 2 days prior to discharge on day 3. On follow-up, the patient denies pain and nausea and enjoys a regular diet without symptoms.

DISCUSSION

The abnormally narrow duodenal lumen as it passes under the SMA may be secondary to an acute angle between the SMA and aorta or a high-riding ligament of Treitz wedging the intersecting segment of duodenum tight into the angle. The angle is between 38 degrees and 65 degrees in normal individuals, but may be well less than 10

degrees in patients with SMA syndrome.⁴ Expressed another way, the distance between the vessels at the level of the duodenum is normally between 10mm to 28 mm,⁵ whereas patients with symptomatic compression have been found to have a mean distance of 6 mm.⁶ Excessive wasting of retroperitoneal fat has also been attributed to the development of this rare syndrome.^{1,6-10} Reports of abnormally low body fat causing SMA syndrome need to be viewed with caution given that the gastrointestinal difficulties of these patients leading to weight loss could blur the cause-effect relationship. If dangerously low body weight were a primary cause, it could be plausibly explained by decreased retroperitoneal fat allowing the small bowel and SMA to lie more posterior, narrowing the vascular angle. Reported cases of successfully reversing symptoms with aggressive caloric augmentation have been attributed to reversing this variable and simultaneously lend credence to the notion that in fact low body weight may be a primary cause.¹¹⁻¹³ The vascular angle may be iatrogenically narrowed after an operation that fixes the SMA into a more posterior position, and this syndrome is reported after ileo-anal anastomosis.¹⁴ Expectedly, because a normal, functional intersection between the duodenum and the vascular angle is dependent upon normal anatomy in 3 dimensions, spinal deformities have been associated with the disease. Particularly, surgical correction of congenital spinal deformities alters the natural position of the aorta via iatrogenic hyperlordosis, thereby narrowing the angle.^{4,15-22} Recent epidemiologic evidence analyzing patients who underwent posterior spinal fusion for scoliosis identified those patients whose weight percentile for height is under 5% to be at risk for developing postoperative SMA syndrome.¹⁵ Paralysis and full-body casts have been associated with the development of acute SMA syndrome in previously asymptomatic patients.²³⁻²⁶ For this reason, SMA syndrome has also been called Cast syndrome. Normally, humans are dynamic beings who continually shift positions. Casted/paralyzed patients may experience duodenal compression by the overlying SMA when they suddenly are subjected to prolonged supine positioning. This notion is supported by the development of SMA syndrome in other types of patients subjected to prolonged supine periods, such as trauma and burn patients.^{11,27} Further, immobilized patients with newly diagnosed SMA syndrome have been successfully treated by simply altering positions.²⁶ Patients with idiopathic SMA syndrome also experience relief from postural changes, although this will not alleviate the disease.^{1,28}

These aforementioned causes provide a more compre-

hendible cause for SMA syndrome. However, as many as 40% of patients have no apparent explainable cause, similar to the case described here.²⁸ In these patients, it is unclear why onset is delayed until early adulthood without evidence of an anatomy-altering event. The great majority of patients will present before age 50.^{29,30} Perhaps natural changes in the GI tract and body habitus with time produce the syndrome in patients fated to have it. Familial cases have been reported including a recent report of monozygotic twins who developed idiopathic SMA syndrome at ages 28 and 29.^{31,32}

Diagnosis currently rests mostly on upper gastrointestinal series and computed tomographic (CT) scans. Features of SMA syndrome on upper gastrointestinal series are a dilated proximal duodenum and vertical or oblique compression of the third portion of the duodenum. Fluoroscopy during the swallow study was highly suggestive in this case with a "to-and-fro" pattern of peristalsis proximal to the obstruction and only small jets of contrast squirting beyond the obstruction. This specific fluoroscopic pattern has been previously described with SMA syndrome,^{33,34} and it appears to offer a strong indication for the presence of the disease. Historically, angiography was recommended.²⁸ However, it currently offers little information that cannot be obtained by CT, it poses significantly greater risks, and it was unnecessary in this case. With appropriate contrast, the CT angiography can be generated in difficult cases with coronal and sagittal reconstruction showing the precise anatomic associations with the vascular angle, the measurement of this angle, and the distance between the vessels.⁴ In our case, abdominal CT with intravenous contrast and upper gastrointestinal series were the only studies required for diagnosis. Although endoscopy is of minor positive diagnostic value, we feel it is mandatory in all patients to rule out intraluminal pathology before either making the diagnosis or initiating treatment. Management of SMA syndrome can be initially conservative with nasogastric decompression, intravenous rehydration, and aggressive nutritional support. Symptoms in immobilized patients first should be considered related to position and treated accordingly. In idiopathic cases similar to ours of patients who are functioning outside of the hospital but are symptomatic, nonoperative options for definitive treatment are not very realistic, and upon confirming the diagnosis, we feel it is appropriate to discuss surgical options.

The first operation performed for SMA syndrome was a duodenojejunostomy in 1908.³⁵ Current surgical options for the disease involve 2 separate concepts: bypass of the obstruction or lysis of the ligament of Treitz. Liberation of

the ligament of Treitz has been historically used with success in small numbers of patients.^{36–38} While this operation is advantageous because enterotomy is avoided, the largest comparative study between the 2 operations demonstrated that 21% of patients failed to respond to lysis of the ligament, whereas all patients treated with bypass experienced resolution of symptoms. Recurrence after ligament liberation occurs simply as a result of post-operative adhesions tethering the bowel into a similar position as the ligament of Treitz had done previously. Recurrence has been described after duodenojejunostomy when the entire anastomosis migrated under the SMA recreating the obstruction, obviously an unusual event.³⁹ Since the introduction of minimally invasive surgery, the results of the open era have thus far been replicated with laparoscopy, albeit in smaller numbers. Laparoscopic lysis of the ligament has been successful in 3 of 4 cases (75%).⁴⁰ There have now been multiple published cases of laparoscopic bypass that have been performed with success.^{34,41–50} Our experience reinforces the idea that laparoscopic duodenojejunostomy should be offered to patients with a diagnosis of SMA syndrome early in the course, because conservative management is unlikely to provide long-term satisfaction.

CONCLUSION

SMA syndrome is a real anatomic clinical pathology resulting in chronic, consistent obstructive symptoms. Diagnosis can be confirmed by upper GI series and CT scan with contrast. Laparoscopic duodenojejunostomy should be considered the treatment of choice for these patients because it offers a high likelihood of an excellent outcome based on the current literature.

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